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OPHTHALMIC SURGEON TO THE PHILADELPHIA HOSPITAL, TO THE CHILDREN'S HOSPITAL, AND TO THE INFIRMARY FOR NERVOUS DISEASES.

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AN EXAMINATION OF THE EYES OF FIFTY CASES OF CHOREA OF CHILDHOOD.

By G. E. DE SCHWEINITZ, M. D. (Univ. of Penn.),
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The cases of chorea from which this study has been made have for the most part been those of patients in attendance at the clinics of Dr. S. Weir Mitchell, Dr. Wharton Sinkler, and Dr. William Osler in the Infirmary for Nervous Diseases. Certain results which should naturally be the outcome of the examinations which follow are not set down, because sufficient time has not yet elapsed to render this possible. The following points have received special attention: The chromatic symmetry or asymmetry of the irides, the development of the face, the condition of the pupils, the acuity of vision, the amplitude of accommodation, the sufficiency or non-sufficiency of the external eye muscles, the state of the refraction, and the appearances of the fundus. A word in regard to the manner in which the cases were studied. Each patient was placed opposite a window and the color of the iris noted, and whether this was alike in each eye or whether differences in color, shade, or tone could be detected. In the same way the two sides of the face and the

development of each orbit were noted. The diameter of the pupils was measured facing a clear sky, and the hour was always the same—between 1 and 2 P. M. For this purpose Randall's pupilometer was employed. The acuity of vision was determined under a uniform illumination at a distance of five metres with Oliver's test-types, each eye, of course, being tried separately. The accommodation was tested with D = 0·5 of Snellen's type and the nearest point of perfectly sharp vision recorded. The muscular relations were studied in the manner especially elaborated by Dr. G. T. Stevens * and the surgeons who have followed his methods. Great care was exercised to have an accurately adjusted trial-frame in which the prisms were set. The object looked at was a point of light against a dark background, placed five metres from the patient. At 30 ctm. or the reading distance the test of v. Graefe was employed, but not the usual coarse dot and line figured in the text-books, but a very fine dot and line, demanding accurate accommodation at the distance employed.

The ophthalmoscopic investigations were all made in the dark room of the infirmary, have in each case been repeated a number of times, and have all been done by the writer himself with Loring's instrument. In a certain number of the cases, twelve in all, a mydriatic correction was made, and hence the findings are accurate. In the others the refraction was measured as nearly as possible in the axis of the eyes. Retinoscopy was often employed as a control test. Fully aware that with complete paralysis of accommodation only can the true refraction of the eye be determined, I am none the less convinced that the findings are reasonably accurate. In this connection I am induced to quote a sentence, which exactly coincides with my own

* "Functional Nervous Diseases, their Causes and their Treatment," by G. T. Stevens, M. D., 1887, p. 194 *et seq.*

view and experience, from Dr. Randall's paper on "A Study of the Eyes of Medical Students" *: "The newer ophthalmoscope of Loring is probably the most perfect instrument in existence for the measurement of refraction; and more than five years' incessant use of it under all circumstances has given the writer a confidence in its readings that work with atropinized eyes and the artificial eye does not show to be overweening. Yet any concealment of its fallibility would be utterly unjust and would tend to weaken the fair claim that this method is the one which, in the absence of complete paralysis of the accommodation, will most frequently approximate accuracy." The ordinary details of the fundus and media of each eye were studied; the appearance of the discs; the absence or presence of the so-called *conus;* the state of the blood-vessels and lymph-sheaths; and the aspect of the retina and chorioid.

Of the fifty patients † examined, ten were males and forty females, or one male to four females. The youngest was a child seven years of age and the oldest twenty-eight. Twelve were under ten years of age, nineteen between ten and fifteen, fifteen between fifteen and twenty, and four twenty years and over. In twenty-five, or 50 per cent. of the cases, the irides were equal in color and shade; in the remaining twenty-five, or 50 per cent., there were differences in shade or tone. In one of these twenty-five only was there any true asymmetry of color. This is interesting in connection with the investigations of Ch.

* "A Study of the Eyes of Medical Students," by Alexander V. Randall, M. D. "Trans. of the State Med. Soc. Penn.," 1885.

† A considerably greater number than this has been examined, but only fifty are recorded, because in these instances the examinations were many times and carefully repeated. The results of the examinations were condensed in tables, but space is lacking to allow of their publication.

Féré.* This observer found asymmetry of color when not dependent on local conditions a very rare symptom. In over six hundred subjects only seven instances were found, and these were imbecile, hysterical, or epileptic. Asymmetry of tone or shade was not rare, especially among the neurotic. In epileptics 26·7 per cent. had this symptom, 62 out of 76 hysterical patients; in choreic patients, those with obstinate facial neuralgia and with sciatica, chromatic asymmetry was observed. When this symptom occurred in the children of the hysterical, Féré believed it was a sign of neurotic tendencies and called for prophylactic treatment. The pupils were of equal size and shape in thirty-nine, or 78 per cent. of the cases, and unequal in eleven, or 22 per cent. of them. The smallest diameter was 2 mm., the largest 6 mm., while the average size of the entire series was 3·39 mm. In twenty-five, or 50 per cent., of the patients the development of the two sides of the face, of the two orbits, and the general appearances of the eyes and their appendages, appeared symmetrical, while in the remaining twenty-five, or 50 per cent., distinct asymmetry in these respects was readily noticeable.

It is well known that the pupils, other things being equal, are larger in young people, while they become smaller and lose both in range and rapidity with advancing years. It is also stated that widely dilated pupils, which react feebly to light, are frequently a feature in cases of chorea (Dr. G. T. Stevens, *op. cit.*, p. 90). Hence it is interesting to observe that in this series the pupillary diameter ranged from 2 mm. to 6 mm., and the average size was only 3·39 mm. While it is true that marked inequality of the pupils is uncommon, except from disease or highly different refraction in

* "Chromatic Asymmetry of the Iris, a Symptom of Neurosis." "Le Progrès médical," September 25, 1885. Abst. in the "Med. News," December 11, 1886.

the two eyes, it is also true that mere differences in the size of the pupils are in no way necessarily a sign of disease. Thus, Iwanow ("The Question of Inequality of the Pupils in Healthy People," "Wratsch," No. 7, 1887, abst. in the "Archiv. f. Ophthalmolog.," Dec., 1887, p. 463) found, in his observations on one hundred and thirty-four healthy young military recruits, equal width of pupil in only twelve. The right pupil was larger in forty-nine, and the left in seventy-three. As Landolt has said ("Examination of the Eyes," trans. by S. M. Burnett, 1879), a lack of symmetry in the face, one half being flatter than the other, would lead us to suspect anisometropia, and all forms of asymmetry of the cranium can cause astigmatism. Individuals with such conformation of features are frequently astigmatic, sometimes anisometropic, the refraction being stronger on the side which is the more fully developed—this eye being myopic or emmetropic, while the other, as a rule, is hypermetropic. Distinct asymmetry existed in 50 per cent. of the cases under consideration; indeed, the proportion was probably higher, as only such cases in which this was readily visible were so classified, and no accurate measures were taken.* Again, however, mere asymmetry of the face must not be looked upon as necessarily a symptom of a diseased state, especially a neuropathic tendency. In Iwanow's series (*loc. cit.*) the face was asymmetrical in one hundred and thirty-one of them, the left being larger in ninety-nine, and the right in thirty-two.

Full acuity of vision—*i. e.*, $\frac{5}{5}$—was present in both eyes in only ten of the cases; in four of them there was normal sharpness of sight in one eye and not the other, while in thirty-two the vision of each eye was less than normal. In

* The relation of this asymmetry to the state of the refraction has not been worked out.

four of the cases it was not possible to record this. In no single instance where it was measured was the amplitude of accommodation equal to the normal standard for the age of the patient.

The state of the refraction in these cases of chorea was the following:

Hypermetropia in twenty-three, or 46 per cent.; hypermetropia in one eye and hypermetropic astigmatism in the other in seven, or 14 per cent.; hypermetropic astigmatism in twelve, or 24 per cent.; myopia in one, or 2 per cent.; myopic astigmatism in three, or 6 per cent.; mixed astigmatism in four, or 8 per cent. In no single instance was a refraction error of less than 1 D found.

As long ago as 1876 Dr. George T. Stevens was attracted by the frequent occurrence of hypermetropia in the eyes of choreic children, and in a paper before the New York Academy of Medicine ("Chorea: its Cause and Treatment," G. T. Stevens, M. D., "Trans. of the N. Y. Academy of Medicine," sec. series, vol. ii, 1876) announced the following proposition: "Chorea is a functional disturbance of the nervous system, which may give rise to organic lesions, and which arises from irritation dependent upon anomalous refraction of the eye, and in a very large proportion of the cases to hypermetropia." In this paper thirty-three cases of chorea are discussed, in which the refraction of the eyes was as follows: Hypermetropia in twenty-four cases, hypermetropic astigmatism in four cases, myopic astigmatism in four cases, unequal myopia in one case.

In the hands of Dr. Stevens the correction of these anomalies of refraction produced excellent results in the curing of the nervous disorder.

About a year later Dr. C. S. Bull analyzed thirty-one cases of chorea with reference to this question, and arrived at very different results ("The Connection between

Chorea and Errors of Refraction," C. S. Bull, M. D., "Med. Rec.," May 26, 1877). Of his patients, fifteen were emmetropic; two were emmetropic in one eye and hypermetropic in the other; eight were hypermetropic of the same degree in each eye; two were hypermetropic of unequal degrees in the two eyes; two were simple hypermetropic astigmatism of the same degree in two eyes; one was hypermetropic in one and simple hypermetropic astigmatism in the other; and one was simple hypermetropic astigmatism in one eye and compound hypermetropic astigmatism in the other. This result, emmetropia in fifteen out of thirty-one cases, is unusual, but must be accepted as correct, as Dr. Bull's patients were examined under atropine. Again, Dr. Ranney ("N. Y. Med. Journal," Jan. 14, 1888), speaking of chorea, says: "I have not found a single case where either manifest or latent hypermetropia did not exist." The writer is certainly in accord with this statement—at least in accord with a statement that anomalies of refraction, in his experience, have always existed, a single case excepted. Dr. G. T. Stevens further classifies in his work on "Functional Nervous Diseases," p. 91, 113 cases of chorea, according to the refraction, as follows: Simple hypermetropia existed in 78; hypermetropic astigmatism in 13; mixed astigmatism in 5; myopia, unequal in the two eyes, in 6; myopic astigmatism in 11.

Adding these various examinations together, we have 227 cases of chorea, of which 212 were ametropic and 15 emmetropic. Of the ametropic cases, hypermetropia or hypermetropic astigmatism was found in 177, or 77·9 per cent.; myopia, or myopic astigmatism, in 26, or 11·5 per cent., and mixed astigmatism in 9, or 3·9 per cent. The remaining 15, or 6·7 per cent., were emmetropic.* This

* It is not evident whether the 113 cases classified in Dr. Stevens's book include the 33 cases in his original paper or not. If they do, then

high percentage of refraction error, especially of hypermetropia and hypermetropic astigmatism, among choreic children is certainly suggestive. It may be interesting to compare these results with the prevailing refraction in children's eyes. To do this, it is only necessary to turn to that most praiseworthy paper by Dr. Randall, "The Refraction of the Human Eye" ("Amer. Jour. of the Med. Sci.," July, 1885), where we find in conclusion No. 2 the following: "Hypermetropia is the enormously preponderating condition in infancy and early childhood, and the first years of school life witness little reduction in its proportion. . . . It was found in 91·26 per cent. of the eyes of infants examined, in 81·75 per cent. of the eyes of young children, and in *76 per cent.* of the 3,358 eyes of children in the elementary school years. So, also, in the higher schools, it constituted at least 56 per cent. of the whole number of eyes studied." Further comment on this point would seem needless, except to point out the singularly close correspondence in the percentage of hypermetropic refraction in the eyes of children in the elementary school years to the percentage of hypermetropia and hypermetropic astigmatism in the eyes of choreic children.

An imperfect equilibrium of the eye muscles was found in every case examined. In a few instances strabismus was present, while in the remainder some form of insufficiency of the ocular muscles was always manifest. Inasmuch as the terms introduced by Dr. Stevens are convenient, they have been employed to designate the muscular defects, which were as follows: Exophoria alone in sixteen, or 32 per cent., of the cases; esophoria alone in thirteen, or 26 per cent., of the cases; hyperphoria alone in one, or 2 per

we have 194 patients in all, of which 149, or 76·7 per cent., had hypermetropia or hypermetropic astigmatism as the existing refraction error. All of the emmetropes were found in Dr. Bull's series.

cent., of the cases; exophoria with hyperphoria in six, or 12 per cent., of the cases; esophoria with hyperphoria in five, or 10 per cent., of the cases; exophoria in accommodation, with esophoria and hyperphoria for the distant point in one, or 2 per cent., of the cases; convergent strabismus in six, or 12 per cent., of the cases; divergent strabismus in two, or 4 per cent., of the cases.

It should be remembered that these were the manifest muscular errors, and have no reference to insufficiencies which may have been latent. It must equally be remembered that, inasmuch as the refraction error was corrected in only a part of these patients, the precaution to examine for muscular defects through the correcting glasses could not nearly always be exercised. What modifying tendency such correction would have had upon the results can only be surmised. In Dr. Bull's series of thirty-one cases (*loc. cit.*) insufficiency of the interni existed in twenty-three. In a considerable number of Dr. Stevens's cases (*loc. cit.;* the percentage is not given) more or less muscular disability was found, and insufficiency of the lateral recti muscles was found in five unassociated with any marked degree of refraction error. The presence of imperfect equipoise of the eye muscles in cases of functional nervous disease, and the belief in some minds that this may be the cause of the disorder, is too well known to need further repetition here. Before deciding the significance of the relation of the high percentage of insufficiency of the ocular muscles in cases of chorea, it may be well to note for a moment its percentage in the eyes of young people generally. In Risley's admirable statistical paper, "Weak Eyes in the Public Schools of Philadelphia" ("Trans. of the State Med. Soc. of Pennsylvania," vol. xiii, 1881), among 554 children of an age ranging from twelve to twenty, and of an average age of seventeen years and one third, insufficiency of the interni was

found in 37·2 per cent. Touching this point, Beselin * has reached some noteworthy results by the examination of 530 cases, of which 363 were Em., 116 H., and 51 M. All having a refraction error of 1 D. or less are called Em. Equipoise in this particular was found in only 140 = 38·5 per cent. of the Em., in 32 = 27·6 per cent. of the H., and in 12 = 23·5 per cent. of the M. Insufficiency of the interni with tendency to divergence for far or for near, or for both, was met in 155 = 42·7 per cent. of the Em., in 52 = 44·8 per cent. of H., and 32 = 62·7 per cent. of M. The contrary condition, with excess of convergence for near or far, was found in Em. 34 = 9·1 per cent., H. 27 = 23·2, and M. 1 = 2 per cent. There were further 2 H. and 1 Em. with divergence for distance and excess of convergence for the near, while of the contrary condition, excessive convergence for distinct and insufficient for near was met in 33 = 9 per cent. Em., 3 = 2·5 per cent. H., and 6 = 11·4 per cent. M. The tests used were a candle-flame for distance and a white Graefe test for the near.

Further comment here is needless, except to point out the large percentage of muscular errors which occur among children who are not and have not been choreic. It is perfectly proper to note that in these school investigations the tests for hyperphoria were apparently not made, and it is to this defect that special attention has recently been drawn.

In regard to the fundus details and the condition of the media, the following points were observed:

In two instances opacities of the media (cornea and vitreous body) were observed. These, of course, bear no reference to the disease. In two, coarse changes in the

* "Untersuchen über Refraction und Grundlinie der Augen und über die dynamischen Verhältnisse der lateral wirkenden Augen-Muskeln an Mädchen von 5–18 Jahren," von Dr. Otto Beselin, "Archiv f. Augenheilkunde," Bd. xiv, zweiter Heft, August, 1884, p. 132.

chorioid were found (atrophic retino-chorioiditis and disseminated chorioiditis), bearing also probably no reference to the chorea. In one case there was slight but evident optic neuritis; suspicion, however, was not lacking that this child had also a brain tumor, although Gowers has several times observed optic neuritis associated with chorea. In one case the discs were greenish and of diminished capillarity, the eyes distinctly amblyopic without corresponding refraction error, but the vessels were unchanged in size. Atrophy of the optic nerve following chorea has been described by William G. Syme *; both sets of vessels were very small, and the trouble was probably due to a former embolus plugging the retinal artery. A similar case is on record by Förster, and Swanzy has seen embolism of the central artery of the retina at the commencement of the attack. Argyll Robertson observed atrophy of one optic disc follow a severe attack of chorea in a young woman. While in the remainder of the cases no perfectly typical, healthy fundus was found, the changes were such as were entirely compatible with systemic health. Quite universally one or another form of conus was present; the veins were frequently distended, often markedly tortuous; coarse retinal striation was quite generally to be found, and the chorioids presented changes ranging from mere woolliness or absorbing interspaces to epithelial chorioiditis. Usually both arterial and venous lymph-sheaths were distended, and many lymph-reflexes were a common feature. In other words, the changes, sometimes slight, sometimes amounting to decided accommodative retinitis and retino-chorioidal disturbances, were such as should be reasonably expected in young, often asthenopic, eyes, suffering, in most instances, with decided errors of refraction. It was, in short, not possible to observe in this

* "Edinburgh Med. Jour.," March, 1888.

series any fundus change that was in any way characteristic of the disease chorea.

The following conclusions seem justifiable:

1. The irides of choreic children quite commonly present chromatic asymmetry in shade, just as the same condition has been found in other forms of nervous disorders.

2. Slight differences in the width of the pupils may be observed, but not more frequently—in fact, not as frequently—as these have been noted in perfectly healthy individuals.

3. Facial asymmetry is present in about one half of the cases, just as this is present in cases of high refractive error, and also in individuals perfectly free from nervous disorders.

4. Hypermetropia and hypermetropic astigmatism are vastly the preponderating conditions of refraction in the eyes of choreic children, being found in about 77 per cent. of the cases, exactly as hypermetropic refraction is the preponderating condition in childhood generally, being found in 76 per cent. of the eyes of children in the elementary school years.

5. Imperfect equipoise of the eye muscles is found in the great majority of the cases, but imperfect equipoise of the eye muscles is very frequently present in the eyes of school-children free from chorea or neuropathic tendencies.

6. Embolism, atrophy of the disc, and optic neuritis may occur during or after attacks of chorea, but appearances in the fundus oculi characteristic of the disease have not been found.

7. As Octavius Sturges remarks: "It seems certain that a fairly constant proportion of chorea is directly connected with what may be called injudicious schooling, . . . but such nice adjustment as shall prevent overstrain on the one hand and overindulgence on the other is practically unattainable." Certainly an endeavor to lessen the overstrain

of the eyes should be made. Hence the refraction errors and muscular defects in these children should be carefully and fully corrected by glasses, by prisms when necessary, or even by judicious surgical interference, and thus a probable exciting element removed; just as we should perform the same service for eyes similarly afflicted in children who are not choreic; just as we should improve the hygiene, remove the anæmia, treat the disabled circulatory apparatus in children who are choreic. Evidence, however, seems quite as lacking that hypermetropic refraction is the basal cause of chorea as it is that the chorea is the cause of the hypermetropia.

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